Original Article

Total Hip Replacement in Sickle Cell Disorder: A Preliminary report of Challenges and Early Outcome of 21 Consecutive Patients

AU Katchy, UE Anyaehie[1], CU Nwadinigwe[1], GO Eyichukwu[1]

Department of Surgery, University of Nigeria Teaching Hospital Enugu, [1]Department of Orthopaedics National Orthopaedic Hospital Enugu, Enugu, Nigeria

Abstract

Objective: The aim of this study is to describe the pattern of presentation osteoarthritic patients with sickle cell disorder (SCD) in our environment, determine the implant sizes taking the peculiar nature of the pathology and our operating environment into consideration, highlight the challenges and technical difficulties encountered during the procedure, measure the functional outcome and observe complications of treatment, recommend ways of improving outcome. **Patients and Methods:** Between November 2008 and November 2012, 29 consecutive primary total hip replacements (THRs) were performed on 21 patients with avascular necrosis of the head of femur secondary to SCD. Patients' evaluation was performed at two different times of follow-up (1 and 5 years, respectively). **Results:** Twenty-one patients were available at 1 year as well as 5-year follow-up. The mean preoperative Harris Hip Score was 20.17 ± 11. The mean postoperative Harris Hip Score was 92.25 ± 13 ($P < 0.001$) at 1 year and 88.75 ± 10 ($P < 0.001$) at 5 years. Eighteen patients had regional anesthesia while three had general anesthesia. The average cup size used was 49.43 with a range of 46–54. The average liner size was 49.43 with a range of 46–54. The head size used in all patients was 28. The average stem size was 6.57 with a range of 6–8. The offset used in all the patients was standard All the patients had 1–2 screw fixation of the acetabular shell. Four patients had complications as follows: periprosthetic fracture 1, superficial wound infection 1, pulmonary complication 1, and abdominal crisis 1. **Conclusion:** THR is a veritable means of treatment of patients with avascular necrosis of the femoral head arising from SCDs. The challenges encountered during the surgery are related to the quality of bone of the affected patient. Arthroplasty Surgeons in our environment must be fully prepared for the challenges by ensuring a preoperative plan that will take care of the technical problems such as recreating femoral canals, wiring of intraoperative fractures as well as treatment of acetabular defects. A detailed planned total hip arthroplasty can be performed in patients with SCD in younger patients with good clinical benefits.

Keywords: *Arthroplasty, challenges, outcome, sickle cell disorder, total hip replacement*

Date of Acceptance:
07-Feb-2018

Introduction

Avascular necrosis of the head of femur is one of the major complications seen in sickle cell disorder (SCD) patients. In terms of pain, level of activity, and function, the hip is one of the most limiting factors in their life.[1,2,3] Patients with SCDs are young

Access this article online

Quick Response Code:

Website: www.njcponline.com

DOI: 10.4103/njcp.njcp_182_17

Address for correspondence: Dr. AU Katchy, Department of Accident and Emergency, University of Nigeria Teaching Hospital, Ituku Ozalla, Enugu, Nigeria.
E-mail: uchekatchy@yahoo.com

How to cite this article: Katchy AU, Anyaehie UE, Nwadinigwe CU, Eyichukwu GO. Total hip replacement in sickle cell disorder: A preliminary report of challenges and early outcome of 21 consecutive patients. Niger J Clin Pract 2018;21:492-5.

and motivated and many of them are at the height of their professional and personal life and could do anything in search of pain relief and restoration of function. In recent years, the treatment of even the most severe forms of this disease has substantially improved with advances in hematologic, visceral, and infectious disease management.[4,5] The lifespan of the patient with SCD has improved with pharmacologic treatments including analgesia, oxygen therapy, hydration, and hydroxyurea[6,7] and can be extended into the sixties.[8,9] Therefore, the treatment of these young patients is particularly important not only for socioeconomic and humanitarian reasons but also because of their life expectancy. In our preliminary report on total hip replacement (THR) in Nigeria, we had reported that a group (7.7%) of our patient population had avascular necrosis of the head of femur from sickle cell disease.[10] In our environment, Alonge and Shokunbi[11] had only presented the use of bipolar arthroplasties and THR as treatment for osteoarthritis in SCD patients while Ekere *et al*.[12] reported the use of THR in two patients. The drawbacks of these studies are that they were presented as case studies without an early outcome assessment. There has been no study in our environment to specifically audit the challenges and early outcome encountered during the treatment of these patients who presented with SCD that had THR. Therefore, the aim of this study is to describe the pattern of presentation in our environment, determine the implant sizes taking the peculiar nature of the pathology and our operating environment into consideration, highlight the challenges and technical difficulties encountered during the procedure, measure the functional outcome, and observe complications of treatment, recommend ways of improving the outcome.

Patients and Methods

Between November 2008 and November 2012, 29 consecutive primary THR was performed on 21 patients with avascular necrosis of the head of femur secondary to SCD. During this period, uncemented THR prosthesis had just been introduced in our institution, National Orthopaedic Hospital Enugu and was commonly used for THR. Patients with osteoarthritis secondary to avascular necrosis from SCD were included in the study. Patients with other indication who are not sicklers were excluded from this study population. The patients were properly evaluated for other SCD complications and medical problems and the average preoperative Packed cell volume (PCV) noted. The preoperative Harris Hip Scores were assessed and the average preoperative Harris Hip Score was 20.17 with a range of 15–30.The average preoperative PCV was 28.43, and the average preoperative transfusion was 1 unit of blood with a range of 0–2units. Only one patient had an exchange blood transfusion few days before surgery after consultation with the hematologist. There was no modification of surgical procedures because of the patient's SCD. The surgeries were carried out under general and regional anesthesia. Routinely, our protocol in the department is regional anesthesia for our lower extremities surgery. However, where epidural injection was not possible due to spinal stenosis general anesthesia was used. We used anterolateral approach for all the patients. The Johnson & Johnson Depuy Corail stems were used for replacement of femoral head and neck, while their Duraloc acetabular components were used for the acetabulum. Fixation of the components was achieved primarily by press-fit technique using screws for acetabular fixation of cup where necessary. There was no remarkable blood loss, and the average loss was 814 ml. The average post up transfusion was less than 2 units of blood. Patients had our normal protocol of rehabilitation comprising; sitting out, walking with Zimmer frame, walking with crutches, and discharged in 2 weeks after removal of staples. The patient had initial postoperative radiographs within 24 hours and subsequent ones taken at the time of assessment of the functional outcome. The post operative Harris Hip Scores were done by the first author at 6 weeks, 3, 6, 12, 24, 36, 48 months, and at 5 years. Our null hypothesis is that patients with SCD will have an improved functional outcome postoperatively.

Results

There were 21 patients with 29 hips; eight patients were bilateral. There were 18 males and 3 females, (M:F = 6:1). The summary on general data of the patient is as shown in Table 1. Eight patients were students, while nine were

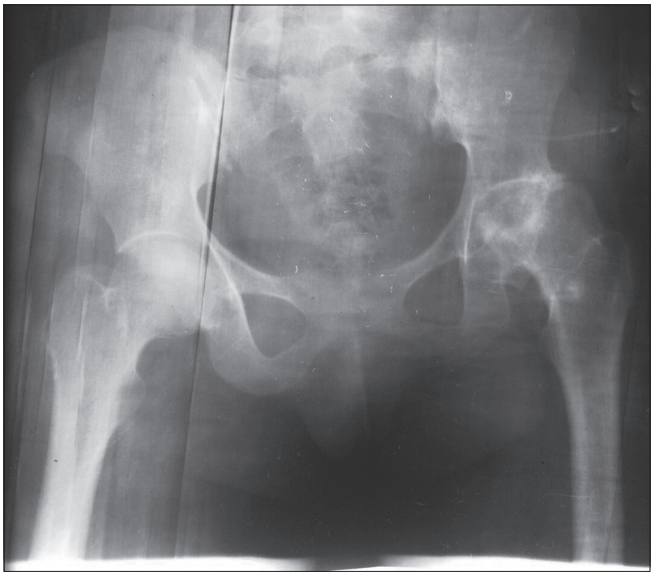

Figure 1: Postoperative X-ray of one of the patients

Table 1: General data

Indices	Frequency
Number of patients	21
Number of hips	29
Bilateral	8 (38)
Males	18 (86)
Females	3 (14)
Male:female ratio	6:1
Mean age of the patients	23.8±3.79
Age range	18-32
Minimum follow-up period	5 years
The mean preoperative Harris Hip Score	20.17±11
Mean postoperative Harris Hip Score (1 year)	92.25±13 (P<0.001)
Mean postoperative Harris Hip Score (5 years)	88.75±10 (P<0.001)
Regional anesthesia (%)	22 (76)
General anesthesia (%)	7 (24)

Table 2: Occupation of patients

Occupation	*n* (%)
Students	8 (38)
Civil servants	9 (43)
Unemployed	4 (19)
Total	21 (100)

Table 3: Implant dimensions

Implants	Dimensions
Cup size	
Mean	49.43
Range	46-54
Liner size	
Mean	49.43
Range	46-54
Head circumference	28
Stem size (range)	6.57 (6-8)
Average number of screws (range)	1.3 (1-2)

Table 4: Complications (n=4; 19%)

Types of complication	*n* (%)
Periprosthetic fracture	1 (25)
Superficial wound infection	1 (25)
Pulmonary complications	1 (25)
Abdominal crisis	1 (25)
Total	4 (100)

civil servants and four were unemployed (see Table 2). All the patients had avascular necrosis of head of the femur as exemplified in Figure 1, with the post operative x-rays shown in Figure 2. The mean preoperative Harris Hip Score score was 20.17. ± 11. The mean postoperative Harris Hip Score was 92.25 ± 13 (P < 0.001) at 1 year and 88.75 ± 10 (P < 0.001) at 5 years. Eighteen patients had regional anesthesia while three had general anesthesia. The average cup size used was 49.43 with a

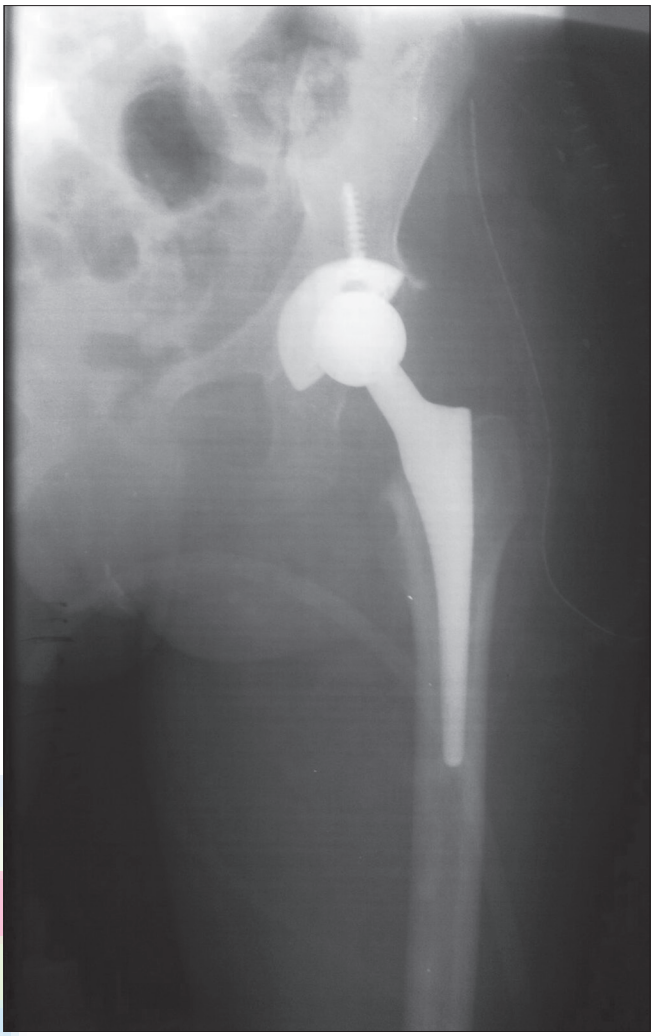

Figure 2: Postoperative X-ray of same patient

range of 46–54. The average liner size was 49.43 with a range of 46–54. The head size used in all patients is 28. The details of the implants used and their dimensions are as shown in Table 3. The offset of all the femoral stems used in all the patients was standard. All the patients had screws. Four patients had complications as shown in Table 4 with periprosthetic fracture 1, superficial wound infection 1, pulmonary complications 1, and abdominal crisis 1. The minimum follow-up period was 5 years.

Discussion

The medical management of patients with SCD continues to improve and this comes with the attendant benefit of patients surviving to adulthood. Most of our patients presented young at the age of thirty two which is within the productive age group. This was further confirmed by the fact that many were students and civil servants who were just beginning to sort out important issues of livelihood. Most patients presented to us with very severe deformity that affected the activities of daily

living as evidenced by their low hip score. We had a male:female ratio of 6:1 which is at variance with the findings of Marulanda *et al*.[1] whose patients presented with a male:female ratio of 1:1.

We observed that for the femoral component the implant size ranged from 6 to 8 in most patients with many of these patients having a size 6 femoral component. The reason for this is the pathological changes of massive fatty bone marrow infiltration and extensive sclerosis that accompanies SCD. The acetabular component sizes ranged from 49 to 54 with an average of 49. Taking our operating environment into consideration and bearing in mind the huge capital outlay involved in arthroplasty, there is need to review the arthroplasty inventory in our national institutions to maintain a reasonable stock of these sizes for patients with SCD as their surgical demand increases. The challenges encountered during the surgery were related to the quality of bone of the affected patient.

We had to recreate femoral canals in most cases to enable the femur take the minimal size of femoral component which was size 6. In some cases, we had fractures while doing this and we had to wire the fractures to ensure healing and femoral component stability. There were also problems of sitting the acetabular component, and we had acetabular defects which we treated as a Type 1 defect.[13]

The functional outcome of our patient improved with postoperative evaluation. The mean Harris Hip Score increased from 20.17. ±11 on admission to 92.25 ± 13 ($P < 0.001$) at 1-year follow-up and 88.75 ± 10 ($P < 0.001$) at 5-year follow-up This is in consonance with the studies of Hickman and Lachiewicz[14] reported 87% good to excellent results in their study of 16 cementless total hip arthroplasty (THAs) in 10 patients with sickle cell disease (mean follow-up, 6 years).

Our complications showed a periprosthetic fracture which we managed as a revision case by wiring. The superficial wound infection and the pulmonary complication were successfully managed. The only abdominal complications resolved.

Acurio and Friedman[15] and Clarke *et al*.[16] with a similar population study and evaluation period had recorded 49% and 59% complications, respectively. This is at variance with our study with a complication rate of 19%. Our results are similar with that of Al-Mousawi *et al*.[17] who recorded a similar complication rate of 17%.

Conclusion

We conclude that THR is a veritable means of treatment for arthritic hip for SCD patients irrespective of their young ages. A detailed planned THA can be performed in patients with SCD with good clinical benefits in our environment.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Marulanda GA, Minniti CP, Ulrich SD, Seyler TM, Mont MA. Perioperative management for orthopaedic patients with sickle cell anaemia. J Orthop Surg (Hong Kong) 2009;17:346-50.
2. Hernigou P, Bachir D, Galacteros F. The natural history of symptomatic osteonecrosis in adults with sickle-cell disease. J Bone Joint Surg Am 2003;85-A: 500-4.
3. Hernigou P, Galacteros F, Bachir D, Goutallier D. Deformities of the hip in adults who have sickle-cell disease and had avascular necrosis in childhood. A natural history of fifty-two patients. J Bone Joint Surg Am 1991;73:81-92.
4. Hernigou P, Habibi A, Bachir D, Galacteros F. The natural history of asymptomatic osteonecrosis of the femoral head in adults with sickle cell disease. J Bone Joint Surg Am 2006;88:2565-72.
5. Steinberg MH. Management of sickle cell disease. N Engl J Med 1999;340:1021-30.
6. Stuart MJ, Nagel RL. Sickle-cell disease. Lancet 2004;364:1343-60.
7. Charache S, Terrin ML, Moore RD, Dover GJ, Barton FB, Eckert SV, *et al.* Effect of hydroxyurea on the frequency of painful crises in sickle cell anemia. Investigators of the Multicenter study of Hydroxyurea in Sickle Cell Anemia. N Engl J Med 1995;332:1317-22.
8. Setty BN, Kulkarni S, Rao AK, Stuart MJ. Fetal hemoglobin in sickle cell disease: Relationship to erythrocyte phosphatidylserine exposure and coagulation activation. Blood 2000;96:1119-24.
9. Platt OS, Brambilla DJ, Rosse WF, Milner PF, Castro O, Steinberg MH, *et al.* Mortality in sickle cell disease. Life expectancy and risk factors for early death. N Engl J Med 1994;330:1639-44.
10. Nwadinigwe CU, Anyaehie UE, Katchy AU. Total hip replacement in Nigeria: A preliminary report. Kuwait Med J 2012;44:291-6.
11. Alonge TO, Shokunbi WA. The choice of arthroplasty for secondary osteoarthritis of the hip joint following avascular necrosis of the femoral head in sicklers. J Natl Med Assoc 2004;96:678-81.
12. Ekere AU, Eze CB, Ebirim LN. A report of two cases of uncemented total hip replacement in patients with sickle cell disease. Niger J Clin Pract 2008;11:257-9.
13. Paprosky WG, Perona PG, Lawrence JM. Acetabular defect classification and surgical reconstruction in revision arthroplasty. A 6-year follow-up evaluation. J Arthroplasty 1994;9:33-44.
14. Hickman JM, Lachiewicz PF. Results and complications of total hip arthroplasties in patients with sickle-cell hemoglobinopathies. Role of cementless components. J Arthroplasty 1997;12:420-5.
15. Acurio MT, Friedman RJ. Hip arthroplasty in patients with sickle-cell haemoglobinopathy. J Bone Joint Surg Br 1992;74:367-71.
16. Clarke HJ, Jinnah RH, Brooker AF, Michaelson JD. Total replacement of the hip for avascular necrosis in sickle cell disease. J Bone Joint Surg Br 1989;71:465-70.
17. Al-Mousawi F, Maliki A, Al-Aradi A, Al-Bagali, Al-Sadadi A, Booz M. Total hip replacement in sickle cell disease. Int Orthop 2002;26:157-61.